A 69-year-old female patient was admitted to our hospital for further cardiac evaluation with symptoms of dyspnea (New York Heart Association class III) and peripheral edema. Her history revealed artificial mitral valve replacement due to mitral stenosis 8 years earlier, severe chronic pulmonary hypertension, and severe tricuspid valve incompetence.

Electrocardiogram showed normofrequent atrial fibrillation. Transthoracic echocardiography indicated moderate biatrial enlargement, although both ventricles had normal dimensions and normal global systolic function. The transmitral prosthetic valve gradient was 14/6 mm Hg. We documented severe tricuspid regurgitation and only mild mitral regurgitation. Nevertheless, echocardiography was difficult to perform because of lack of an adequate acoustic window and metal artifacts of the mitral valve prosthesis. Reflections at the epicardial surface close to the mitral valve were therefore interpreted as calcifications of pericardial layer, and restrictive pericarditis was expected to be the most likely cause of the patient’s symptoms and diastolic left ventricle dysfunction.

By right heart catheterization, severe postcapillary hypertension was confirmed (systolic/diastolic/mean pulmonary artery pressure, 65/23/43 mm Hg; mean pulmonary wedge pressure, 29 mm Hg). In all cardiac cavities, an end-diastolic pressure equilibration was measured (Figure 1). Pressure curves did not show a dip-plateau phenomenon. Therefore, constrictive pericarditis was excluded. However, in a 64-row multidetector computed tomography scan, the endocardial layer of the left atrium (LA) and the aortic arch were heavily calcified (Figure 2), whereas the pericardium itself did not show any relevant calcifications. As an additional finding, the patient showed tracheobronchopathia osteoplastica (Figure 2A and 2B). Cardiac MRI was performed to further assess ventricular and atrial function (Figure 3). Any acute or persistent cardiac inflammation or interstitial storage cardiomyopathy could be excluded by edema-sensitive T2-weighted short T1 inversion recovery (STIR) images and T1-weighted images early and late after intravenous Gd-DTPA according to the protocol previously described. The
dynamic cine MRI study showed no relevant changes in LA volumes during the heart cycle, indicating a stiff LA due to the heavily isolated calcification of the endocardial surface of the LA (Figure 3).

The patient’s symptoms were explained as combination of elevated LA pressure and pulmonary hypertension. The singular calcification of the stiff LA led most likely to reduced filling of the LA itself. In combination with diastolic dysfunction and former mitral stenosis, it might be the main cause for backward failure and secondary pulmonary hypertension, tricuspid incompetence, and right heart failure. Therapeutic management included medication for diastolic dysfunction and decrease of afterload with diuretics (hydrochlorothiazide, furosemide), angiotensin-converting enzyme inhibitors and β-blockers. A surgical approach was abandoned because of low probability of success.

**Discussion**

Two rare conditions were found in 1 patient: a calcified LA and a tracheobronchopathia osteoplastica. The presence of LA calcification, also known as “porcelain atrium” or “coconut atrium,” has been reported as a result of extensive rheumatic pancarditis. The etiology of tracheobronchopathia osteoplastica, a mostly asymptomatic condition of the tracheobronchial tree with a probably benign course, is still unknown. Several other mechanisms may account for endo-
cardial LA calcification, including postoperative complication after mitral valve replacement due to hematoma and inflammation; atrial wall response to chronic strain forces present in the setting of mitral disease, idiopathic, or in the context of rheumatic fever in combination with tracheobronchopathia osteoplastica and calcification of the aorta; ischemic myocardial damage with dystrophic calcification in remodeling; or hypercalcemia and hyperphosphatemia (eg, in metastatic calcification or in combination with renal failure). In our patient, malignancies, electrolyte disturbances, or renal failure could be excluded. Although the patient was unable to recall a history of rheumatic disease, which may have been oligosymptomatic, it seems to be the most likely diagnosis.

Figure 3. Cine MRI of a 4-chamber view of the heart during diastole (A) and systole (B), showing contraction of the ventricles and differences in size of the right atrium but not of the isolated calcified LA. Dephasing artifacts at the tricuspid valve indicate tricuspid incompetence. Metal artifacts are obvious at the artificial mitral valve.

Disclosures
None.

References
Endocardial Calcification of Left Atrium, Tracheobronchopathia Osteoplastica, and Calcified Aortic Arch in a Patient With Dyspnea
Ulrike M. Müller, Stephan Gielen, Gerhard C. Schuler and Matthias Gutberlet

Circ Heart Fail. 2008;1:290-292
doi: 10.1161/CIRCHEARTFAILURE.108.799437

Circulation: Heart Failure is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 2008 American Heart Association, Inc. All rights reserved.
Print ISSN: 1941-3289. Online ISSN: 1941-3297

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circheartfailure.ahajournals.org/content/1/4/290

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in Circulation: Heart Failure can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to Circulation: Heart Failure is online at:
http://circheartfailure.ahajournals.org//subscriptions/